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PC-06

Assessment of Right Ventricular Myocardial Fibrosis Using Two-Dimensional and Three-Dimensional Speckle-Tracking Echocardiography in Patients with End-Stage Heart Failure

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Background: Myocardial fibrosis (MF) is a common finding in patients with advanced heart failure (HF) irrespective of the etiology of cardiomyopathy and can play an important role in affecting ventricular function in these patients. Previous studies show that right ventricular (RV) free wall longitudinal strain (LS) assessed by 2-dimensional speckle-tracking echocardiography (2D-STE) correlates with the extent of RV MF. 3-dimensional speckle-tracking echocardiography (3D-STE) has emerged as a more accurate tool for assessing the RV function than 2D-STE. We aimed to identify which RVLS is the most reliable parameter to predict RV MF in patients with end-stage HF. **Methods:** 105 consecutive patients with end-stage HF undergoing heart transplantation were enrolled in our study. The conventional RV function parameters (RV ejection fraction, RV fractional area change, tricuspid annular plane systolic excursion, and tricuspid annular plane systolic velocity), 2D-LS and 3D-LS of RV free wall were obtained in these patients. The degree of MF was quantified by using Masson trichrome stain in RV myocardial samples. The study population was divided into 3 groups according to the degree of MF on histology (mild, moderate, and severe MF). **Results:** Patients with severe MF had lower 3D-LS, 2D-LS and conventional parameters of RV function compared with those with mild and moderate MF. RV MF strongly correlated with 3D-LS ($r = 0.73$; $p < 0.001$), modestly with 2D-LS ($r = 0.53$; $p < 0.001$), weakly with conventional RV function parameters ($r = -0.49$ to -0.29 ; $p < 0.01$). 3D-LS correlated best with the degree of MF ($r = 0.73$ vs 0.29 to 0.53 ; $p < 0.05$) compared with 2D-LS and conventional parameters. 3D-LS had the highest accuracy for detecting severe MF (area under the curve 0.899 vs 0.804; $P = 0.0187$) compared with 2D-LS. The model using 3D-LS ($R^2 = 0.61$; $p < 0.001$) was found to predict the degree of MF most accurately than that with 2D-LS ($R^2 = 0.46$; $p < 0.001$) and the RV fractional area change ($R^2 = 0.44$; $p < 0.001$) value. **Conclusion:** RV 3D-LS may be the most accurate functional measure that predict the extent of RV MF in patients with advanced heart failure.

Rate of Change of Ventricular Strain and Strain Rate Following Fontan Operation					
Variable	Mean (Standard Deviation)	Rate of Change per Year since Fontan Operation (95% CI)	P-value	Effect of Composite Outcome on Rate of Change per Year since Fontan Operation (95% CI)	P-value
Global Longitudinal Strain (%)	-19.06 (4.19)	0.03 (-0.10, 0.16)	0.63	-0.15 (-0.60, 0.30)	0.51
Global Longitudinal Strain Rate (1/s)	-0.959 (0.29)	0.011 (0.002, 0.02)	0.02	-0.015 (-0.045, 0.016)	0.35
Free Wall Longitudinal Strain (%)	-20.41 (4.91)	-0.03 (-0.78, 0.11)	0.64	-0.27 (-0.80, 0.26)	0.32
Free Wall Longitudinal Strain Rate (1/s)	-1.05 (0.33)	0.007 (-0.004, 0.017)	0.23	-0.015 (-0.051, 0.020)	0.40
Global Circumferential Strain (%)	-20.64 (4.41)	-0.11 (-0.25, 0.03)	0.14	-0.19 (-0.71, 0.33)	0.48
Global Circumferential Strain Rate (1/s)	-1.0 (0.29)	0.014 (0.003, 0.024)	0.01	0.031 (-0.003, 0.024)	0.07

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Strain Rate Worsens in Long Term Longitudinal Follow Up of Patients Following Fontan Operation

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Background: Qualitative assessment of ventricular function in single ventricle heart disease is hindered by a lack reproducibility. Recent studies have explored strain and strain rate as tools for qualitative assessment, but experience with these tools is limited. We sought to investigate longitudinal changes in strain and strain rate following Fontan operation. **Methods:** A retrospective observational study of a Fontan inception cohort at Children's Hospital of Philadelphia (CHOP) 2006-2007. Echocardiograms obtained from inception (2006-2007) through 2017 were identified. Those with a minimum of four echocardiograms and six months between each echocardiogram were included. Change in strain and strain rate were assessed across the cohort and then compared between those who did and did not meet a composite outcome of protein losing enteropathy (PLE), plastic bronchitis, transplant or death. **Results:** There were 286 echocardiograms from 2006-2017 in 38 unique patients (76% male, 37% hypoplastic left heart syndrome (HLHS)) for an average 7.5 echocardiograms per patient (range 4-11). Six of the 38 patients met the composite outcome (3 PLE, one plastic bronchitis, one transplant and one death). Global longitudinal strain rate increased (worsened) by 0.011 s⁻¹/year ($p = 0.02$) and global circumferential strain rate increased (worsened) by 0.014 s⁻¹/year ($p = 0.01$) over the study period. The cohort mean values for global longitudinal, free wall and global circumferential strain were all worse than what would be expected for a healthy age-matched population without heart disease, but did not have a statistically significantly change over time. There was no difference in the rate of change of the strain parameters in those who met the composite endpoint versus those who did not. The rate of change in the strain characteristics did not differ based on ventricular morphology. **Conclusion:** This study demonstrates a decline in global longitudinal and circumferential strain rate over a decade of follow-up after Fontan. These may be early signs of cardiac dysfunction that pre-date qualitative signs of deterioration. The lack of difference between those who reached the composite endpoint and those who did not may be an issue of statistical power, or may be due to the multifactorial nature of failure of the Fontan circulation.

PC-08

Evaluation of Myocardial Work under Different Loading Conditions: A True Marker of Contractility?

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Background: Myocardial strain obtained by speckle tracking echocardiography is a sensitive parameter of systolic function and has established prognostic value. However, one of its major limitations is load dependency, which can lead to the misinterpretation of myocardial function in case of increased afterload. By incorporating both myocardial deformation and afterload, myocardial work (MW) can overcome this limitation, and it reflects contractility presumably better than strain. Accordingly, we aimed to verify myocardial work as a marker of contractility in three separate rat models with different loading conditions. **Methods:** Three rat models were used: (1) pressure overload-induced heart failure (evoked by transverse aortic constriction [TAC], $n = 12$ [+12 sham]), (2) volume overload-induced heart failure (evoked by abdominal aortic fistula [AAF], $n = 12$ [+12 sham]), and (3) exercise-induced physiological hypertrophy (EX, induced by 200 mins/day swimming for 12 weeks, $n = 10$ [+10 controls]). Speckle tracking echocardiography was performed to assess global longitudinal strain (GLS), which was followed by pressure-volume analysis to assess left ventricular pressure and to compute cardiac contractility (preload recruitable stroke work [PRSW]). Global MW index (GMWI) was assessed by calculating the area of the left ventricular pressure-strain loop. **Results:** GLS was found to be preserved in the ACF group (-14 ± 3 vs. $-15 \pm 2\%$, $p = NS$) whereas it was decreased in TAC (-7 ± 3 vs. $-13 \pm 1\%$, $p < 0.001$) and increased in EX (-21 ± 4 vs. $-17 \pm 2\%$, $p < 0.05$) compared to their corresponding age-matched sham/control groups. PRSW and GMWI were markedly decreased in ACF compared to sham (59 ± 14 vs. 111 ± 40 mmHg, $p < 0.001$; 1365 ± 444 vs. 1863 ± 218 mmHg%, $p < 0.01$), however, they did not differ between TAC and its sham group (112 ± 68 vs. 115 ± 34 mmHg; 1657 ± 675 vs. 1603 ± 229 mmHg%, both $p = NS$). The EX group was characterized by increased values of PRSW and GMWI (148 ± 27 vs. 108 ± 12 mmHg, $p < 0.001$; 2973 ± 672 vs. 2248 ± 375 mmHg%, $p < 0.01$). GLS showed weak correlation with PRSW ($r = -0.363$, $p < 0.01$), whereas GMWI correlated moderately with

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PRSW ($r=0.613$, $p<0.001$) in the entire study cohort. The correlation between GMW1 and PRSW remained significant in all three models when analyzed separately; nonetheless, GLS correlated with PRSW only in EX. **Conclusions:** Under different loading conditions, MW reflects contractility better than GLS, and it represents a promising novel marker of myocardial function.

PC-09

The Role of Screening Fetal Echocardiogram after the Prenatal Detection of Extracardiac Malformations

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Background: Congenital heart disease (CHD) is increasingly detected prenatally and up to 50% of fetuses with CHD have an extracardiac malformation (ECM). It is unknown whether fetuses diagnosed with an ECM should undergo routine screening fetal echocardiography for CHD. We aimed to investigate the utility of screening fetal echocardiography among patients diagnosed with any type of ECM at a single multidisciplinary fetal center. **Methods:** We performed a retrospective review of all patients presenting for fetal echocardiography due to an ECM at Columbia University's Center for Prenatal Pediatrics between January 2013 and December 2018. We noted both the type and number of ECM (increased nuchal thickness/cystic hygroma, single umbilical artery, and agenesis of the ductus venosus were excluded), type of CHD, presence of prenatal genetic diagnosis, and whether the CHD was detected on obstetric anatomy sonogram. Univariable analyses, including correlation and chi-square, were performed. **Results:** Among 646 patients with at least one ECM, 75 (11.6%) had CHD diagnosed at a mean gestational age (GA) of 25.6 ± 0.2 weeks. The most common forms of CHD were ventricular septal defect (65.3%), coarctation (37.3%), and tetralogy of Fallot (14.7%). The frequency of CHD by type of ECM was 30.6% for craniofacial, 23.5% for musculoskeletal or limb anomalies, 16.4% for central nervous system, 15.1% for gastrointestinal, 12.6% for genitourinary, 10.3% for arteriovenous malformations, and 9.8% for thoracic. The mean number of fetal ECMs was 1.32 ± 0.03 ; an increase in the number of fetal ECMs correlated with likelihood of CHD ($r=0.98$, $p<0.0001$). Of fetuses with ECM and a prenatal genetic diagnosis, 36.1% had CHD as compared to 10.2% of those without a genetic diagnosis ($p<0.0001$). Nearly half, 48%, of the CHD in this cohort was detected on obstetric anatomy sonogram at a mean GA of 23.9 ± 0.2 weeks. **Conclusion:** CHD was diagnosed commonly among fetuses with any type of ECM. Since CHD may not always be detected on obstetric sonogram but may impact decision-making and perinatal care, all patients with a diagnosis of ECM should undergo routine screening fetal echocardiography.

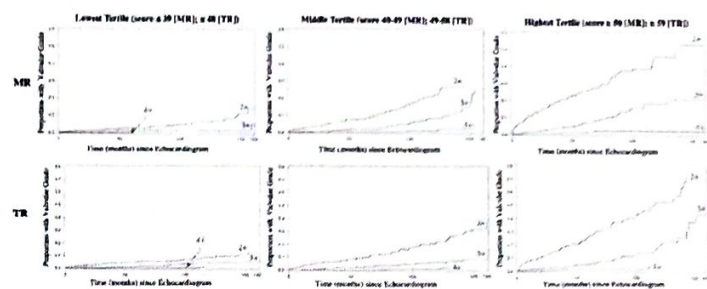
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Development and Validation of an Echocardiographic Prediction Rule for Long-Term Progression of Mitral and Tricuspid Regurgitation

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Background: Progression of mitral (MR) and tricuspid (TR) regurgitation on echocardiography (TTE) demonstrates wide inter-individual variability. Individualized prediction of progression is needed. **Methods:** Structured TTE reports from 38,908 patients with 1-3+ MR or TR at our institution, 2000-2017, were used to model MR and TR progression ($\geq +1$ increase in severity) separately, using specialized proportional hazards models (to adjust for interval- and left-censored data). Estimates, derived in a random 2/3 sample, were used to create a progression score, tested in a 1/3 leave-out sample. **Results:** 5,379 (15.4%) patients with MR and 3,630 (13.2%) with TR had progression at a median [IQR] 909 [240-2226] days (MR) or 917 [224-2160] days (TR). Derivation c-statistics were 0.76 (MR) (95% CI 0.75-0.78) and 0.76 (TR) (95% CI 0.74-0.77). MR and TR score variables were similar and included age, left atrial size, TR gradient, peak aortic velocity, LVEF, and initial MR/TR severity. The highest score tertile had greater progression (MR: 20.1%; TR: 21.2%) than the lowest tertile (MR: 3.3%; TR: 4.4%; $p < 0.001$) (Figure). Validation c-statistics were 0.76 (MR) (95% CI 0.74-0.79) and 0.74 (TR) (95% CI 0.71-0.76). Predicted progression was similar to observed. Using a threshold of 10% progression, the low-score tertile progressed over >205 months, the intermediate-score tertile over 104-121 months, and the high-score tertile over 39-58 months. **Conclusions:** A simple rule using routinely collected echocardiographic variables predicts long-term MR/TR progression with very good accuracy. This rule may be used to personalize TTE surveillance intervals for MR/TR, but should be externally validated. Using a threshold of 10% progression, current TTE surveillance guidelines are too frequent for individuals with a low and intermediate score.

Figure: Cumulative Incidence Functions Demonstrating the Proportion of Individuals with Each Valvular Severity Grade at Time Intervals since the Initial Echocardiogram Demonstrating Mild (1+) Mitral or Tricuspid Regurgitation, Stratified by Tertile of Valvular Progression Score



The X-axis indicates the proportion of individuals with each valvular severity grade (1+, 2+, 3+, 4+) since the initial echocardiogram demonstrating 1+ MR (top panels) or TR (bottom panels) stratified by valvular progression score tertile. The proportion with 2+ MR/TR is indicated by the blue line, 3+ by the red line, and 4+ by the green line. Survival are determined at the time of the initial echocardiogram. Estimates are right-censored at the last observation. Separate curves demonstrating progression in those with 2+ or 3+ MR/TR at baseline by score tertile are not shown.

PC-11

Consensus-Based Development of a Pediatric Echocardiography Complexity Score - Design, Rationale, and Preliminary Results of a Quality Improvement Collaborative

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Background: Current models of stratification of complexity in congenital heart disease are primarily based on the surgical palliation and associated outcomes. The utility of these models in the non-surgical setting has not been evaluated despite the increased availability of guidelines for standardized diagnostic testing and imaging-based quality improvement (QI) initiatives. We report on the development and early phase of validation of a pediatric echocardiography complexity (PEC) score. **Methods:** The American College of Cardiology's Quality Network assembled a panel of pediatric echocardiographers from 12 centers to develop, by consensus, the PEC scoring tool. Using a previously published echocardiographic scoring algorithm to categorize complexity based on the lesion and/or indication, the panel a) refined the complexity categories, b) included modifiers that add complexity to the performance of an echocardiogram, and c) identified potential applications of the PEC score. Each center used the PEC score tool on 15 random inpatient and outpatient echocardiograms, and the group discussed possible opportunities to enhance the usability of the tool. **Results:** The PEC scoring tool is shown in Figure 1a. Among the 180 studies submitted, 42% were outpatient, 64% follow-up, 20% pre-operative, and 11% were sedated studies. The distribution of PEC scores for all studies are shown in Figure 1b. Studies in the PEC 5 category involved the most heterogeneous diagnoses with a median scan time of 38 (13-447) min. Of the 27 studies that required additional scanning by senior sonographer or cardiologist, 78% were in the PEC 4 and 5 categories. Potential complicating modifiers suggested for addition to the tool included presence of tracheostomy, known genetic syndromes, and unrepaired/repaired lesions. **Conclusions:** The preliminary results of this multicenter study suggest that the PEC scoring tool is feasible and easily applicable in a variety of clinical settings. The modifiers included in the study design add further value in characterizing study complexity. The PEC tool can be utilized as a clinical and a research tool for correlation with diagnostic errors, allocation of resources, and assessment of physician and sonographer effort in performing, interpreting, and teaching echocardiograms.

